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What Happens in the Formation of Ureteral Duplication? A Case Report and Brief Literature Review

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Abstract

Radiologists frequently diagnose two ureteral duplications including complete and incomplete types. Incomplete ureter duplication, which is also called bifid ureter, refers to a situation in which the ureteric bud bifurcates before meeting the metanephric blastema. On the other hand, complete ureter duplication, which is also named double ureter, defines another situation in which two separate ureteric buds, on either side of the embryonic body, are formed from the mesonephric duct (Wolffian duct) and then arrive in the metanephric blastema. Therefore, two separate ureters, renal pelvis, as well as a duplex kidney (upper and lower poles) are formed in a common renal capsula. Embryologically, in complete ureter duplication, the ureteric buds rotate 108 degrees when incorporating into the urogenital sinus, which is known as Weigert-Meyer rule. In this posture, the upper and lower poles of the kidney are drained by the laterocranial and mediocaudal orifice, respectively. Complete duplication is more common in women than men and is often associated with vesicoureteral reflux, ectopic ureterocele, and/or ectopic ureteral insertion. In the present case report, a 40-year-old woman, mainly complaining of abdominal and lower back pain, was discovered to have bilateral complete ureteral duplication on computed tomography (CT) scan, as well as kidney, ureter, and bladder x-ray radiography. Moreover, the embryological causes were discussed in forming the duplicated ureter and duplex collecting system.

Keywords: Ureteral Duplication, Congenital Anomalies, Computed Tomography Scan, KUB Radiography, Embryology

Introduction

Embryologically, during gastrulation (week 3 of gestational), three types of mesoderm, namely, the paraxial, intermediate, and lateral plate mesoderm are formed on both sides of the midline of the embryonic body (1,2). The intermediate mesoderm, which is also called the nephrotome, forms several parts in the body such as gonads, a portion of adrenal glands, reproductive system, and nephric structures, that is, the pronephric, mesonephric, and metanephric kidneys, weeks 4 and 5 of gestational, in the cranial-to-caudal sequence, and ureters (1,2). At the fourth week of gestation, mesonephric ducts (Wolffian ducts) develop on either side of the body. Then, they grow toward the lower lumbar region in order to form a part of the posterior wall of the definitive bladder when fusing to anterolateral (ventrolateral) walls of the cloaca at week 4 of gestation, forming the ureteric buds (metanephrogenic diverticulum) at week 5 of gestation (1,2). After the formation of the definitive kidney (metanephros) at the fifth week of gestation, an interaction between metanephric mesenchyme (blastema) and the ureteric bud results in forming different parts including ureters, as well as collecting duct systems such as the renal pelvis, major and minor calyces. In addition, such interaction further leads to collecting ducts and tubules from a single ureteric bud, as well as glomerulus, capsule, and nephron tubules which are originated from the metanephric tissue (1-4), the details of which are illustrated in Figure 1A. It means that the ureteric bud fails to bifurcate unless it enters into the metanephric blastema. Therefore, a bifid ureter or a Y-shaped bifid ureter, which is also called incomplete duplication, is formed if the ureteric bud bifurcates before meeting the metanephric blastemal Figure 1B displays the formation of the above-mentioned ureter (2-4). On the other hand, a ureter with complete duplication is formed if two separate ureteric buds are formed from the mesonephric duct on one side of the embryonic body. As a scientific term, duplex kidney (i.e., duplex or duplicated collecting system) describes a situation in which two separated pelvicalyceal systems drain a single renal parenchyma into the incomplete (partial) (i.e., 2 ureters are drained into

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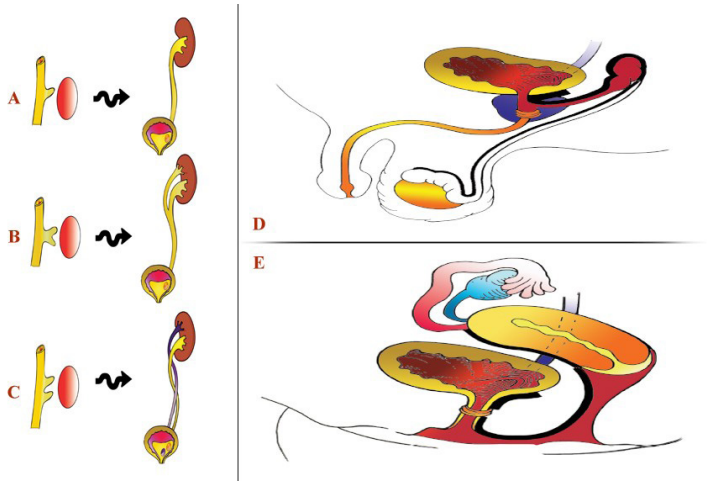

Figure 1. (A) Normal situation (single system). A single ureteric bud sprout from the Wolffian (mesonephric) duct prior meeting with metanephros to form a single ureter and renal collecting system. (B) Abnormal situation (duplex system). A ureteric bud bifurcates before meeting with metanephros. Therefore, although two collecting systems form, there is a common distal ureteral orifice (incomplete duplication). (C) Abnormal situation (duplex system). Two separated ureteric buds sprout from the Wolffian (mesonephric) duct that meet the metanephros one by one. Accordingly, two complete duplications of the ureter (i.e., duplex or duplicated ureter) and renal collecting system occurs. The lower pole ureter (yellow), which deranges the lower pole of the kidney, is analogue with the single-system ureter while the upper pole ureter (purple), which deranges the upper pole of the kidney, is ectopic (Weigert–Meyer rule) and inferomedially related to another ureteral orifice inserted to the bladder. (D) The ectopic pathways of the upper pole ureter in males and in the situation of complete ureteral duplication. The pathway of the upper pole ectopic ureter is outlined in dark. The *thick dark line* shows the most common ectopic locations which are from the bladder Trigone to the bladder base and external sphincter; *medium dark line* illustrates the less common ectopic locations in the ejaculatory duct or seminal vesicle; *thin dark line* displays the rare ectopic location in the vas deferens. (E) The ectopic pathways of the upper pole ureter in females and in the situation of complete ureteral duplication. The pathway of the upper pole ectopic ureter is outlined in dark. *Thick dark line* shows the most common ectopic locations which are from the bladder Trigone to the bladder base and external sphincter; *medium dark line* depicts the less common ectopic locations into the urethra below the external sphincter, perineum and/or vagina; *thin dark line* represents the rare ectopic locations into the uterus and/or fallopian tube.

the bladder by a single common ureter) and/or complete (i.e., double ureters that drain separately into the urinary bladder) ureteral duplication (Figure 1C) (2-4). In the complete ureteral duplication situation, two ureters drain separately into the bladder or one ureter opens into the bladder and another opens into genital tracts such as the vagina, the urethra, or the vulval vestibule (vestibulum vagina); this process is depicted in Figures 1D and E (2-4). Anatomically, the ureters are two long tubular structures (25-30 cm in length and 3-4 mm in diameter) with thick muscular walls and a narrow lumen, which extend from renal pelvis to the urinary bladder on either side of the body (5). In the urinary tract, the duplication of the ureter is considered as the most anatomical anomaly, which occurs in approximately 0.7%-4% of the population while rising to 8% in the United States and is more common in females than males (4,6). Based on some reports, incomplete duplication may be associated with other congenital anomalies and defects (4,6).

The current study presented a case report of bilateral complete duplication of the ureters which was diagnosed using a three-dimensional computed tomography (CT) urography.

Case Presentation

A 40-year-old woman referred to Alavi hospital of Ardabil (Ardabil, Iran) complaining of a 2-year abdominal and lower back pain which had been intensified over the past 2 months. The patient neither reported a history of diabetes mellitus, hypertension, asthma, smoking, drug abuse, drug allergy, or fever in the past nor had she any urinary problem including infection and frequent urination. The spiral CT scan of the abdomen and pelvis with intravenous contrast of the patient was requested by the urologist. Blood urea nitrogen level and creatinine test were completely normal before performing the CT scan (urea=37`, creatinine=0.9). The CT scan of abdominopelvic with IV contrast was performed at intervals of 60 seconds after the injection in order to examine renal parenchyma and check the kidney and urethra secretory phase 15 minutes after the injection. In addition, three-dimensional reconstruction of images was implemented after completing the CT scan. Moreover, KUB radiography (kidney, ureter, and bladder x-ray) was performed and both kidneys were found to have two separate ureters (Figure 2).

Discussion

Ureteral duplication or duplex (duplicated) collecting system is considered as the most common anatomical anomalies of the urinary tract which may be found in association with other genitourinary tract anomalies and defects (4). According to the reports, complete duplication is more common in women compared to men and is frequently associated with vesicoureteral reflux, ectopic ureterocele, and/or ectopic ureteral insertion while bifid ureter is often related to the uretero-ureteral reflux or ureteropelvic junction obstruction of the lower pole of the kidney (4). On the other hand, the four

most common complications of complete duplication include ureteropelvic junction obstruction of the lower pole, ectopic ureteral opening, ectopic ureterocele, and vesicoureteral reflux (4,7). Based on the reports of a study, a duplicated collecting system occurs unilaterally approximately four times more than bilaterally (7).

As previously mentioned, an interaction between the ureteric bud and metanephros induces signaling pathways which result in forming renal collecting systems and nephrons. Therefore, any failure in the signaling pathways leads to urinary tract anomalies. For instance, the overexpression of the glial cell-derived neurotrophic factor-RET was reported to induce the formation of multiple ureteral buds (2).

However, two medical terms, namely, upper or lower pole ureter and upper and lower poles of a duplex kidney, are used for describing the above-mentioned situations. For instance, if a mesonephric duct, which is sprouted as two ureteric buds (i.e., cranial and caudal), or a mesonephric duct, which is divided into two ureteric buds (Y-shape, i.e., cranial and caudal protrusion), penetrates inside metanephric blastema, then the cranial and caudal buds induce a signal which leads to the formation of the cranial and caudal poles of the kidney, respectively, each of which has its own renal pelvis and ureter (1-4).

Additionally, the caudal ureteric bud normally arrives into the bladder wall (Trigone area) when the mesonephric duct is absorbed into the posterior wall of the urinary bladder on either side of the embryonic body (1). On the other hand, the cranial ureteric bud attaches to the caudal ureteric bud in the situation of incomplete duplication or bifid ureter, then it opens into the Trigone of the bladder by a single duct (1). Moreover, in the situation of complete duplication, the cranial bud forms a caudal ectopic ureter which drains the cranial pole of the kidney while the cranial bud, which drains the caudal pole of the kidney, forms the normal ureter that connects to the bladder (1,3,4). Based on the Weigert-Meyer rule, the ectopic and normal ureters cross to each other, then the ectopic ureter can open into different sites in a male or female (1,3,4,7).

In males (Figure 2D), an ectopic ureter can open into the base of the bladder and external sphincter (most common), the ejaculatory duct or the seminal vesicle (less common), and the vas deferens (rare) while, in females (Figure 2E), it can open into the base of the bladder and external sphincter (most common), the urethra or the vagina (less common), as well as the uterus or the fallopian (uterine) tube (rare). Unlike males, the ectopic ureter can open into the proximal or distal to the external urethral sphincter in females, therefore, women are always wet because of the dripping from the ectopic ureter (1, 4).

Further, medical imaging techniques such as

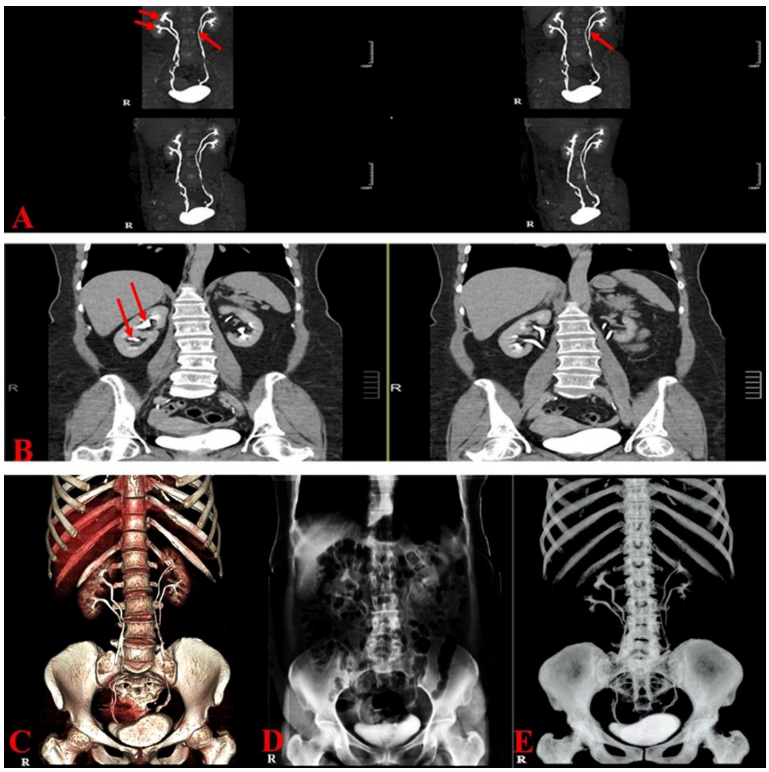

Figure 2. (A) Three-dimensional reconstruction of the ureter in different directions. (B) computed tomography scan with intravascular contrast in secretion phase (after 15 minutes). (C) Three-dimensional reconstruction. (D) Kidney, ureter, and bladder X-ray imaging. (E) Three-dimensional reconstruction of the abdominopelvic with contrast in the negative view.
Note. The arrows show two separate pelvicalyceal collecting systems and double ureters.

ultrasonography, voiding cystourethrography, radionuclide cystography, intravenous pyelography, dimercaptosuccinic acid renal cortical scintigraphy, CT, magnetic resonance urography, dynamic renal scintigraphy, and diuresis renography can be used as diagnosing methods for renal collecting system duplication or complete duplication of the ureters (4, 8).

In general, the current study only discussed the causes related to the formation of complete ureteral duplication. A thorough knowledge of anatomical variations in the human body such as duplicated collecting system is very important for the surgeons in order to avoid any accidental traumatic injury of the ureter during hysterectomy surgery or any other therapeutic surgeries (4, 10-12). Moreover, radiologists should be aware of the ureter variations in order to have a correct interpretation of the radiographs.

Conflict of Interests

Authors have no conflict of interests.

Ethical Issues

The present case report was approved by the Local Ethics Committee of Tabriz University of Medical Sciences.

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